



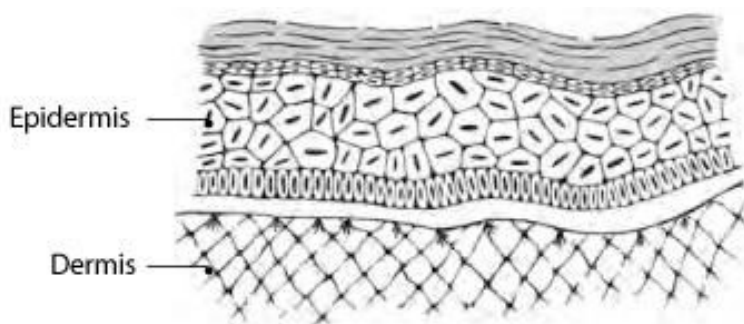
## What is it?

### Points To Remember About Pemphigus

- Pemphigus causes blisters on the skin and in the mouth, nose, throat, eyes, and genitals.
- The type of pemphigus depends on where the blisters form.
- Most people with pemphigus can control it with medicines. These medicines can have side effects, so talk to your doctor about all medicines you are taking.
- See your doctor if you have long-lasting blisters on the skin or mouth, so that you can be tested for pemphigus.

Pemphigus is a type of disease where the body's immune system attacks healthy cells in the top layer of skin (epidermis). It causes blisters on the skin and in the mouth, nose, throat, eyes, and genitals.

Some forms of pemphigus can cause death if not treated.



Skin structure

## Who gets it?

Pemphigus affects people of all races and cultures. Groups at higher risk are:

- People of Mediterranean descent.
- Eastern European Jews.
- People who live in the rainforests of Brazil.

Men and women both get pemphigus at the same rate. It is most common in middle-aged and older adults. But it can occur in young adults and children.

## What are the types?

The type of pemphigus depends on where the blisters form.

- **Pemphigus vulgaris** is the most common type of pemphigus in the United States. Most cases start with mouth blisters. The blisters can be painful but usually do not itch or leave scars.
- **Pemphigus foliaceus** most often starts with sores or blisters on the face and scalp. Blisters then show up on the chest and back. It can also cause loose, moist scales on the skin. Most of the sores are itchy, but not painful. This type of pemphigus does not cause mouth blisters.
- **Pemphigus vegetans** causes thick sores in the groin and under the arms.
- **IgA pemphigus** is the least harmful type. The blisters look like those in pemphigus foliaceus. This type can also cause small bumps with pus inside. This type of pemphigus is caused by an antibody called IgA.
- **Paraneoplastic pemphigus** is a rare type of pemphigus. Special tests may be needed to diagnose it. It occurs in people with some types of cancer and can lead to:
  - Painful mouth and lip sores
  - Cuts and scars on the lining of the eyes and eyelids
  - Skin blisters
  - Severe lung problems.

Pemphigoid is a disease like pemphigus. Pemphigoid leads to deep blisters that do not break easily. Pemphigoid is most common in older adults and may cause death.

## What are the symptoms?

Pemphigus causes blisters on the skin and in the mouth, nose, throat, eyes, and genitals.

## What causes it?

Your immune system makes antibodies, which attack viruses and harmful bacteria. With pemphigus, antibodies instead attack healthy cells in the skin or mucous membranes. As a result,

- Skin cells separate from each other.
- Fluid collects between skin layers.
- Blisters form and may cover a large area of skin.

The cause of this immune system attack is not known. Pemphigus does not spread from person to person. It does not appear to be passed from parent to child. But some people's genes put them more at risk for pemphigus.

## **Is there a test?**

Early diagnosis is important, so see your doctor if you have long-lasting blisters on the skin or mouth. Pemphigus is rare, and your doctor may test for other diseases first.

The doctor should do all of these tests. No single test is right all the time.

- A complete history and physical exam.
- Remove a sample of a blister and look at with a microscope.
- Do chemical tests on a skin sample to determine the type of pemphigus.
- A blood sample to measure pemphigus antibody levels.

## **How is it treated?**

Pemphigus is treated with medicines that:

- Control the immune system.
- Treat infections.

These medicines can have major side effects. Ask your doctor to tell you about them. Tell each doctor you see about the medicines you take.

## **Who treats it?**

Doctors who treat pemphigus include:

- Dermatologists, who treat skin problems.
- Dentists, who can tell you how to take care of your gums and teeth if you have blisters in your mouth.

## Living With It

Pemphigus is rarely fatal. Most people with pemphigus can control it with medicines. But pemphigus and its treatments can cause:

- Lost time at work.
- Weight loss.
- Loss of sleep.
- Stress.

Support groups can help you cope with the disease.

## For More Info

### **U.S. Food and Drug Administration**

Toll free: 888-INFO-FDA (888-463-6332)

Website: <https://www.fda.gov>

Drugs@FDA at <https://www.accessdata.fda.gov/scripts/cder/daf>. Drugs@FDA is a searchable catalog of FDA-approved drug products.

### **Centers for Disease Control and Prevention, National Center for Health Statistics**

Website: <https://www.cdc.gov/nchs>

### **American Academy of Dermatology**

Website: <https://www.aad.org>

### **The International Pemphigus Foundation**

Website: <https://www.pemphigus.org>

### **American Autoimmune Related Diseases Association, Inc.**

Website: <https://www.aarda.org>

### **National Organization for Rare Disorders**

Website: <https://www.rarediseases.org>

If you need more information about available resources in your language or other languages,

please visit our webpages below or contact the NIAMS Information Clearinghouse at [NIAMSInfo@mail.nih.gov](mailto:NIAMSInfo@mail.nih.gov).

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